Gallbladder agenesis: A rare anomaly of the biliary system

Safra kesesi agenezi; safra yollarının nadir görülen bir anomalisi

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Dear Editor;

A 44-year-old man visited our outpatient clinic for a routine checkup with no complaint. He was not using any medications or alcohol. His vital signs and physical examination were normal. Liver enzymes were all within normal range, but cholestatic enzymes were mildly elevated: alkaline phosphatase: 124 (30-120 U/L), gamma-glutamyl transferase: 81 (0-55 U/L), and total bilirubin: 1.10 (0.3-1.2 mg/dL). Hepatobiliary ultrasonography (US) revealed a liver of slightly elevated size (16 cm) with normal structure and parenchymal echogenicity. Intra- and extra-hepatic bile ducts were not dilated. US did not visualize the gallbladder clearly (Figure 1). The patient was anxious when he learned that US showed an invisible gallbladder. Two months later, he visited our outpatient clinic again. He was still anxious. We learned that he sought medical attention at other hospitals, and repeated US revealed

an invisible gallbladder, but doctors suspected a contracted or porcelain gallbladder. Computer tomography was performed, which confirmed the absence of a visible gallbladder with normal intra- and extra-hepatic bile ducts similar to suspected contracted or porcelain gallbladder. We performed magnetic resonance cholangiopancreatography (MRCP), which revealed normal intra- and extra-hepatic bile ducts and pancreatic duct with no dilation or filling defect. The gallbladder was not detected (Figure 2).

Gallbladder agenesis (GA) is a rare congenital anomaly of the biliary system with an estimated incidence of 13-65 individuals per 100 000 cases (1). Three groups of GA were identified in 1988 (2): asymptomatic (35%), which comprised individuals diagnosed at autopsy or laparo-

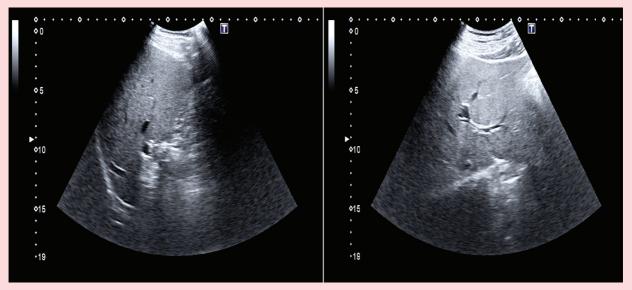


Figure 1. Sonography of the biliary system demonstrating undilated intra- and extra-hepatic bile ducts and no evidence of the gallbladder.

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Uyaroğlu OA. Gallbladder agenesis: A rare anomaly of the biliary system. The Turkish Journal of Academic Gastroenterology 2020;19:38-39. **DOI: 10.17941/agd.705807**

Manuscript received: 12.05.2019 • Accepted: 20.09.2019

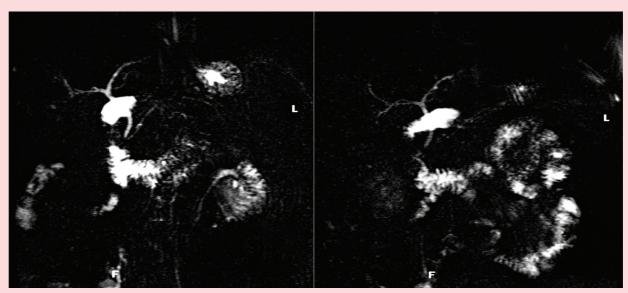


Figure 2. Magnetic resonance cholangiopancreatography: absence of the gallbladder.

tomy; symptomatic (50%), whose symptoms were chronic right upper quadrant pain (90%), dyspepsia (30%), nausea/vomiting (66%), fatty food intolerance (37%), and jaundice (35%); and foetal anomalies (15%), which indicated congenital anomalies incompatible with life. The most common clinical presentation is right upper quadrant pain mimicking biliary colic. Most cases of symptomatic GA are scheduled for surgery with a misdiagnosis of cholecystitis or choledocholithiasis. If the diagnosis of GA is made intraoperatively, patients often are exposed to complications from prolonged exploration. Aborting the procedure rather than complete further exploration is recommended (3). US is the most sensitive technique for imaging the biliary system. Most cases of GA have been reported as "contracted/fibrotic gallbladder" on

ultrasound. The main method to establish GA is MRCP, which is a noninvasive and well-demonstrated imaging method in the evaluation of the biliary tract. It can also demonstrate an excluded and/or ectopic gallbladder (4,5).

MRCP enables the correct diagnosis to be made by a noninvasive examination, thereby suppressing our patient's anxiety. The patient was informed about his congenital malformation to avoid possible unnecessary surgical exploration in the future. GA should be considered whenever the gallbladder is not visualized in routine imaging methods in patients with or without symptoms to reduce misdiagnosis and unnecessary surgeries.

"The authors declared that there is no conflict of interest regarding the publication of this article."

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